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## What is post-polio syndrome (PPS)?

*[EDITOR'S NOTE: SOMETIMES WE NEED TO GET BACK TO BASICS. WHILE THOSE OF YOU WHO HAVE BEEN AWARE OF PPS FOR SOME TIME MAY BE A BIT BORED BY THIS ARTICLE, WE HAVE MANY NEWCOMERS IN OUR GROUP WHO DESPERATELY NEED INFORMATION ABOUT WHAT'S HAPPENING TO THEM. THIS ARTICLE WAS TAKEN FROM THE WEB SITE OF GAZETTE INTERNATIONAL NETWORKING INSTITUTE ([HTTP://WWW.POST-POLIO.ORG/](http://www.post-polio.org/)). G.I.N.I., AS IT IS CALLED, IS PERHAPS THE DE FACTO AUTHORITY ON POST-POLIO SYNDROME.]*

Post-polio syndrome is a condition that affects polio survivors years after their recovery from their initial bout with polio. An interval of 30 to 40 years usually elapses before the first PPS symptoms occur, but intervals as short as 8 years and as long as 71 years have been documented.

*What are the most common symptoms of PPS?*

Fatigue, new weakness, and pain are the most common symptoms of PPS, but many other symptoms can also occur, including:

- Fatigue, which occurs in 60% to 90% of people with PPS, may be the most common symptom, and is probably most responsible for disability associated with the syndrome. Some people experience PPS-related fatigue as general fatigue, which often is described as a flu-like exhaustion that worsens as the day progresses. This type of fatigue can also increase during physical activity, and may cause difficulty with concentration and memory. Others experience muscle fatigue, a form of muscle weak-

ness that increases with exercise and improves with rest. In addition, many individuals experience symptoms of both general fatigue and muscle fatigue

- New weakness usually occurs in muscles that were affected during the original polio infection; however, this weakness also can occur in muscles that were not affected by the original polio infection, or at least did not seem to be affected at the time
- People with PPS may experience pain in their muscles and/or joints. Muscle pain is usually felt as an aching or sore feeling that occurs after light physical activity, and it frequently improves with rest. Joint pain can be chronic (lasting for a long time) or intermittent (occurring off and on for short periods of time), and it may intensify during physical activity
- Less common PPS symptoms may include:
  - new muscle atrophy (loss of muscle bulk)
  - respiratory insufficiency (breathing difficulties)
  - dysarthria (speech disturbances)
  - dysphagia (difficulty swallowing)
  - muscle cramps
  - cold intolerance
  - fasciculations (muscle twitches)
  - joint deformities

*How many people have PPS?*

It is estimated that there are currently 640,000 survivors of polio in the United States. Studies suggest that 20% to 40% of those who have recovered from the initial polio infection go on to develop PPS; thus, there may be as many as 128,000 to 256,000 individuals with PPS in the United States.

*How does PPS affect quality of life?*

PPS is not typically a life-threatening condition, but it may cause significant discomfort and disability.

The most common disability caused by PPS is deterioration of mobility. People with PPS may also experience difficulties performing daily activities such as cooking, cleaning, shopping, and driving. Assistive devices such as canes, crutches, walkers, wheelchairs, or electric scooters may be necessary for some people. If symptoms are extremely severe, these individuals may be required to change occupations or stop working altogether.

#### *How does PPS cause these symptoms?*

The exact mechanism by which PPS causes fatigue, pain, and new weakness is not completely understood, but it is thought to be related to the gradual loss of individual nerve cells that make contact with muscle fibers, and subsequent loss of nerve transmission to these fibers. During the original polio infection, the poliovirus attacks motor neurons (nerve cells responsible for muscle movement), causing injury or death of those nerve cells. If this process occurs on a large enough scale, it can result in loss of muscle function, including weakness or paralysis. However, the body often employs strategies to compensate for lost functions -- in this case, surviving nerve cells sprout extra branches, called axonal sprouts, that are able to attach to muscle fibers that have lost their original nerve supply. After this process of nerve reattachment, or reinnervation, the muscle fibers are able to work again, sometimes as well as before. Some researchers have suggested that PPS develops because these extra axonal sprouts cannot "hold" forever, but instead get weaker over time due to "overexertion" -- in other words, they cannot sustain this additional activity indefinitely, because they were not designed to do so. Eventually, the sprouts degenerate, and the muscle fiber again loses its ability to contract.

#### *How is PPS diagnosed?*

It may be difficult to obtain a proper diagnosis of PPS, in part because there are no definitive tests for this condition. Some doctors dismiss PPS symptoms as being caused by psychological conditions such as depression. In addition, common PPS symptoms such as fatigue, pain, and new weakness may also be caused by many other diseases. As a result, the diagnosis of PPS is one of exclusion, meaning that a doctor can only diagnose PPS

by eliminating other potential causes of the patient's symptoms. In general, a doctor may suspect PPS in a polio survivor if:

- The individual is suffering from new muscle weakness, generalized or muscular fatigue, or pain involving the muscles and/or joints
- The individual has experienced at least 10 years of good health (i.e., muscle functions have not deteriorated) since recovering from polio

If these criteria are met, a doctor must still make sure that the symptoms are not caused by other medical conditions, including pulmonary (lung), cardiac (heart), hematologic (blood), or endocrine (hormonal) diseases, cancer, or some chronic infections. In addition, musculoskeletal diseases such as fibromyalgia, rheumatoid arthritis, and osteoarthritis may also be confused with PPS. Fibromyalgia, a common musculoskeletal disorder that can cause generalized pain and tenderness as well as fatigue, is frequently seen in polio survivors, and may sometimes co-exist with PPS. In addition, many neurologic diseases -- including amyotrophic lateral sclerosis (Lou Gehrig's disease), multiple sclerosis, and Parkinson's disease -- may cause PPS-like symptoms. However, even if an individual is found to have another condition that causes PPS-like symptoms, this does not rule out the possibility that he or she may also have PPS.

#### *Is there a cure for PPS?*

There is currently no cure for PPS. However, several drugs are being studied in the hopes that they will provide relief from some of its symptoms (see below). In addition, people with PPS have a wide range of non-drug options for symptom management. Depending on the individual, a typical strategy may involve physical therapy with careful use of exercise; occupational therapy, including instruction on lifestyle changes, such as avoiding overexertion; use of assistive devices such as canes or walkers; appropriate weight loss if needed; and use of over-the-counter or prescription pain medications. Often, these management strategies are developed in consultation with the individual's primary care physician, along with a team of other healthcare professionals that may include a physiatrist (a doctor specializing in rehabilitative medicine), a neurologist, a rheumatologist or orthopedist, a physical therapist, an occupational therapist,

a speech therapist, an orthotist (a person who creates orthopedic devices), a social worker, and a dietitian.

### *How to manage new weakness?*

Strengthening, stretching, and aerobic exercises may be useful for the management of new weakness, as is appropriate weight loss. However, not everyone benefits from the same types of exercise, and some individuals should avoid exercise altogether. In general, people with PPS should avoid exercise that causes joint pain, and should not exercise to the point of muscle pain or fatigue. It is important to avoid muscle overuse because this may result in increased weakness. If muscle overuse does occur, rest or supportive devices such as braces may be beneficial. Assistive devices -- such as canes, crutches, manual wheelchairs, electric wheelchairs, and motorized scooters -- can be helpful for severe muscle weakness.

### *How to manage fatigue?*

One way to manage PPS-related fatigue is through the use of energy conservation techniques. For example, daily activities that are not considered "critical," such as making a bed, can be discontinued. Or, household items and supplies can be relocated to make them more accessible. Some people may consider certain lifestyle changes, such as working only part-time or taking a job that is less physically demanding. In addition, it helps to learn to pace daily activities, which means taking regular breaks during long periods of physical activity. Regular rest periods or daytime naps, especially in the early afternoon, can be beneficial for managing fatigue. (People with PPS who work should also consider this type of rest schedule.) Finally, it is important to get a good night's sleep; doctors can provide advice on how to improve the quantity and quality of sleep. (It should be noted that breathing disorders such as sleep apnea may interfere with sleep and contribute to fatigue.)

### *How to manage pain?*

There are different types of pain associated with PPS, and management strategies must be tailored accordingly. Some types of pain may be of muscular origin -- such as muscle cramps or fibromyalgia

-- while others may result from joint and soft tissue problems that are secondary to PPS, including osteoarthritis, tendinitis, ligament strains, and joint deformities. One way that people with PPS can manage pain is to modify the use of their extremities -- for example, by periodically resting a leg that is causing discomfort. Certain "physiotherapeutic" techniques, including the application of ice or superficial heat, as well as the use of ultrasound and a method called transcutaneous electrical nerve stimulation, or TENS, may be beneficial for pain management. Strengthening exercises, supportive devices such as braces, and assistive devices can also help to relieve pain. Acetaminophen (e.g., Tylenol) and nonsteroidal anti-inflammatory drugs (such as ibuprofen [e.g., Advil], naproxen sodium [e.g., Naprelan, Aleve], and etodolac [Lodine]) are effective pain relievers that can be obtained over-the-counter or by prescription. In rare cases, steroid injections or surgery may be required to relieve a patient's pain.

### *How are other PPS symptoms managed?*

An estimated 13 percent of people with PPS develop new breathing difficulties, most commonly caused by weakness of the respiratory muscles. Other factors may be pulmonary (lung) disease; spinal deformities such as scoliosis or kyphosis; obesity; smoking; and/or sleep-associated breathing disorders (e.g., sleep apnea, a common problem in polio survivors). Many polio survivors who used a ventilator during their original polio infection may need to return to the use of a ventilator. Management of breathing and sleep problems can be achieved largely through the use of nocturnal (nighttime) ventilation. A small percentage of people with PPS suffer from dysphagia, or difficulty swallowing (often caused by weakness of the muscles of the pharynx or larynx). A videofluoroscopic evaluation may be needed. Swallowing problems can be managed with a variety of techniques, including eating smaller and more frequent meals during the day, avoiding eating when fatigued, and turning the head to one side or tucking down the chin while swallowing.

### *Is PPS associated with emotional problems?*

Many individuals have difficulties adjusting to new disabilities. For some people with PPS, reliving

their childhood experiences with polio can be a traumatic and even terrifying experience. To make matters worse, many doctors know very little about PPS, and some believe that the condition is only in the patient's head. Lacking proper medical and psychological advice, people with PPS are often forced to depend on their own resources to help them cope with their condition. Fortunately, PPS is gaining increasing attention in the medical community, and there is a growing number of healthcare professionals who understand PPS and can provide appropriate medical and psychological help. In addition, there are PPS support groups, newsletters, and educational networks that provide up-to-date information about PPS while offering individuals the knowledge that they are not alone in their struggle (see contact information below).

#### *Are there any promising new treatments for PPS?*

Several drugs are being studied in the hopes that they may prove effective in relieving some of the symptoms of PPS:

- Human growth hormone (Humatrope) has been found to increase the production of a hormone in the body -- insulin-like growth factor I or IGF-1 -- that stimulates nerve cells to sprout additional extensions (axons). This drug, which is already approved for other uses, was tested in 6 people with PPS, producing little or no improvement in the subjects' muscle strength, endurance, or recovery after fatigue. However, the study investigators suggested that longer-term treatment with human growth hormone might help some individuals with PPS
- Bromocriptine mesylate (Parlodel), a drug that is used to treat conditions such as Parkinson's disease, was found to improve symptoms of fatigue in 3 out of a total of 5 persons with PPS. Researchers are hoping to conduct larger studies to confirm the

benefits of this drug

- Recombinant human insulin-like growth factor I (rhIGF-1) was shown to enhance the recovery of strength in people with PPS after they engaged in tiring exercises, but the drug did not alleviate excessive fatigue caused by the exercises. The study involved 22 people with PPS
- Pyridostigmine (Mestinon), a drug currently approved to treat muscle fatigue associated with the neuromuscular disorder myasthenia gravis, has shown some promise in the treatment of PPS. In one study of the drug involving 27 people with PPS, more than half of the subjects reported less fatigue after receiving pyridostigmine. A separate study involving 27 people with PPS found that patients taking pyridostigmine showed minimal improvements in strength (based on objective measures) and fatigue levels (based on the individual's own opinion) The potential benefits of pyridostigmine are being investigated further in a clinical trial called the North American Postpoliomyelitis Pyridostigmine Study, or NAPPS. Preliminary results have been released and pyridostigmine was not found to provide significant benefits with respect to quality of life, fatigue, or isometric muscle strength compared with placebo, although a trend was noted towards increased strength in very weak muscles. [ For more detailed information, see Talk Back ] This trial of 126 people with PPS is designed to determine the effectiveness of pyridostigmine in relieving PPS symptoms using measures of fatigue, muscle strength, and general health. NAPPS will also determine the incidence and severity of any side effects associated with pyridostigmine. Adverse effects were seen in approximately 50% of individuals in previous trials of pyridostigmine; these side effects included loose stools and intestinal cramps, blurred vision, increased urinary frequency, muscle cramps, and fasciculations (muscle twitches).

### Patient checklist

Check off the items in the following list that apply to you. If you check off the first item on this list and any of the subsequent ones, see a doctor.

✓	I have had polio in the past and I now have:
	unaccustomed fatigue (this may be "general fatigue"), a feeling of total exhaustion, or "muscle fatigue", a rapid tiring of the muscles that increases with exercise and improves with rest
	new muscle weakness (may include muscles originally affected by polio infection, as well as muscles seemingly unaffected at the time)
	pain in my muscles and/or joints
	breathing or swallowing difficulties
	increased sensitivity to cold temperatures
	a decline in my ability to perform basic daily activities, such as climbing up stairs or bathing
	difficulty sleeping

For further information on polio and PPS, contact the International Polio Network, (IPN), an organization with a forty-year history of collecting, creating, and disseminating information on issues relating to polio and post-polio syndrome.

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Some of us were joking at the last meeting about how we could blame everything on PPS (remember the comment about the 105 year old lady who died of post-polio syndrome?)

Penny Jensen clipped this cartoon out. We couldn't think of an appropriate PPS caption, but you get the idea.

